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The value of video-EEG monitoring to diagnose juvenile myoclonic epilepsy

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ABSTRACT

Objective: A diagnostic accuracy of conventional electroencephalography (EEG) is approximately 50% at best. We aimed to determine the accuracy of video-EEG monitoring (VEM) for a correct diagnosis and the feasibility of its clinical application. The data from all 55 patients (M:F = 31:24) with juvenile myoclonic epilepsy (JME) who underwent VEM were reviewed according to the clinical history, brain imaging and video-EEG findings.

Results: Age at seizure onset ranged from 10 to 25 (15.5 ± 2.7 years). The age at VEM ranged from 15 to 46 (21.8 ± 5.8 years) and 57% (29/51) showed seizures. Of those, 20 patients (69%) showed myoclonic jerks alone, whereas 3 (10%) showed generalized seizures alone. Both of these conditions were observed in 6 patients (21%). Interictal abnormalities alone without clinical seizures were detected in 16 patients (31%). Atypical semiologies such as asymmetric myoclonus or versive seizures were observed in 18 patients (35%) during video monitoring. Interestingly three patients complained of visual aura on history. The duration of VEM ranged from 1 to 6 days (1.8 ± 1.1). Overall, 88% of patients showed an EEG abnormality with/without seizure, concordant with JME. Among 10 patients with a normal conventional EEG before VEM, 9 showed interictal or ictal EEG abnormalities during approximately 1-day of VEM.

Conclusions: VEM for 1 or 2 days is appropriate for making a correct diagnosis of JME, especially in patients having an atypical semiology and a normal result on the conventional EEG.

1. Introduction

Juvenile myoclonic epilepsy (JME) is one of the idiopathic generalized epilepsy syndromes, classically characterized by three semiological features: (1) morning myoclonic jerks in both arms, (2) a brief interruption of consciousness and (3) generalized tonic–clonic seizures. The clinical diagnosis is supported by compatible electroencephalography (EEG) changes, which show generalized polyspike-and-wave patterns without background slow waves, normal brain imaging and a normal cognitive function.[1] Although JME is relatively common and is clearly defined (as listed above),[2–4] it is commonly mis- or under-diagnosed due to the strong reliance on these "typical" three semiologies and EEG results. Simply based on history from patients alone at outpatient clinic, we may regard myoclonic jerking in only one limb[5] and brief unresponsiveness[6] as focal motor and complex partial seizure, respectively. Depending on result of EEG alone to diagnose JME may be troublesome. It is well known that an entirely normal EEG as well as focal or lateralized epileptiform discharges[7–10] can make a diagnosis difficult. Several studies[1,5,11–13] have suggested that even repetitive EEGs do not reveal the suggestive abnormalities of generalized epilepsy in 21–54% of patients. Moreover, accurate diagnosis can be more difficult when we obtain an unreliable or atypical seizure history alone.

Video-EEG monitoring (VEM) system is widely applied to describe the seizure semiology and localization of the seizure focus. In patients with JME, it is held that VEM can demonstrate myoclonic jerks and absence seizures, which might go unrecognized by a patient or doctor; it can also be applicable to detect the typical EEG findings of patients with JME more easily. There have been some observations[1,5,14] that VEM is often helpful to clarify an atypical semiology and electrographical findings, but to date the literature regarding the diagnostic value of VEM in patients with JME has been scarce. Previous studies did not include cases that did not demonstrate clinical seizures and placed more emphasis on ictal recording and semiology. The aim of this study was to determine the feasibility of VEM for making an early and correct diagnosis of JME and if there is a patient subset for which VEM is better indicated.

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2. Materials and methods

VEM was performed on 1935 patients in our center from January 1995 to April 2006. Among them, 55 patients were diagnosed with JME during the entire follow-up period and we included all such patients who underwent VEM in this study. There were 31 men and 24 women, whose ages ranged from 15 to 46 years at VEM (mean 21.8 ± 5.8 years). A diagnosis of JME was made according to the criteria of International League Against Epilepsy.[15] The diagnosis was confirmed from each patient's historical or videotaped semiology, ictal or interictal EEGs, brain imaging and drug response to antiepileptic drugs (AEDs) during the follow-up period. The patients' medical records were reviewed regarding their history, and two epileptologists analyzed the video-EEGs until they reached a consensus. The typical EEG changes compatible with a diagnosis of JME included: (1) a symmetrical generalized polyspike-and-wave, (2) a symmetrical generalized isolated spike-and-wave, and (3) symmetrical generalized spike-and-wave complexes at 3 Hz or over 3 Hz (fast type).[13] Atypical findings such as irregular bursts of slow waves without a discernible generalized epileptiform discharge were not considered to be a characteristic EEG feature of JME.

2.1. Conventional EEG at outpatient clinic

Conventional EEGs (c-EEGs) were performed according to the international 10–20 system at the EEG laboratory in the same hospital without any modification of the patient's AED. Each EEG session lasted for 30 min and any EEG changes occurring during hyperventilation and photic stimulation were also observed.

2.2. Brain imaging

Magnetic resonance imaging (MRI) was performed on 31 patients, and computer tomography (CT) was performed on one. The standard MRI technique was as described.[16] In some cases, 3-mm thick sections of the tentative symptomatogenic regions were also obtained considering the patient's c-EEG and semiology.

2.3. Video-EEG monitoring

In our center, 458 patients were diagnosed as JME from January 1995 to April 2006, of whom 55 patients (12%) performed VEM. Interictal and ictal EEGs and clinical seizures were recorded using a VEM system with the electrodes placed according to the international 10–20 system, including the additional anterior temporal electrodes. VEM was performed for the following reasons: (1) to confirm a diagnosis of JME in patients with drug resistance or a discrepancy between their clinical diagnosis and the c-EEG results performed at an outpatient clinic; (2) to clarify the epilepsy syndrome of patients referred from other centers with an equivocal classification; (3) to identify any vague semiology; or (4) for the first diagnosis of any untreated patient.

VEM was performed after withdrawing the AEDs with the exception of phenobarbital. The EEGs were reviewed using the bipolar and referential montages. All patients were routinely advised not to go to sleep earlier than usual and additional provocations such as photic stimulation or hyperventilation were administered in the morning only to those patients not showing ictal or interictal epileptiform discharges during the previous night. In general, VEM was stopped when the typical EEG pattern of generalized epilepsy or seizures compatible with a diagnosis JME occurred. Occasionally, the length of monitoring was extended when there were inconsistent findings.

2.4. Seizure outcome

All patients included in our study had follow-up duration of at least 1 year after VEM. Their seizure outcomes were measured at last follow-up.

2.5. Statistical analysis

One-way ANOVA was used to examine the statistical significance of any difference between the continuous variables using SPSS for Windows (Version 12.0).

3. Results

Table 1 summarized the patient's clinical data and VEM results. The age of seizure onset ranged from 10 to 25 years (mean 15.5 ± 2.7) and the mean duration between seizure onset and VEM was 6.2 ± 5.8 years. The mean follow-up duration since VEM ranged 12–197 months (mean 57.3 ± 47.2).

3.1. Seizure history

The prior seizure history was taken from a witness or the patients themselves. Among the 55 patients, 41 (75%) had suffered both myoclonic jerks and generalized seizures. Of these, 11 (20%) experienced all three types of seizures, myoclonic jerks, generalized seizure and brief loss of consciousness; two (4%) had had myoclonic jerks plus brief losses of consciousness and one patient (2%) had had only myoclonic jerks.

Table 1
Summary of clinical characteristics and video-EEG results

	No. (%)	Median VEM duration (day) (range)
Seizure history		
M + G	41 (75)	1 (1–6)
M + G + A	11 (20)	2 (1–4)
M + A	2 (4)	2 (2)
Only M	1 (2)	4 (4)
Initial impression before VEM		
JME	36 (65)	1 (1–3)
JME plus	12 (22)	2 (1–6)
Others	7 (13)	3 (1–4)
Lesion on MRI		
Yes	6 (19)	2 (1–6)
No	26[a] (81)	1 (1–5)
Monitored seizures		
Typical[b]	11 (22)	1 (1–5)
Atypical	18 (35)	2 (1–4)
No seizure	22 (43)	1 (1–6)
Interictal EEG abnormality		
Typical[c]	43 (84)	1 (1–5)
Atypical	4 (8)	1.5 (1–3)
Normal	4 (8)	1 (1–6)
Ictal EEG abnormality		
Typical[c]	25 (83)	1 (1–4)
Atypical	5 (17)	2 (1–5)

VEM, video-EEG monitoring; M, myoclonic jerk; G, generalized seizure; A, absence seizure.
[a] Includes one patient undergoing CT.
[b] Denotes a symmetrically involved myoclonic jerk or generalized seizure.
[c] Denotes a symmetrical generalized polyspike-and-wave, a symmetrical generalized isolated spike-and-wave or symmetrical generalized spike-and-wave complexes at 3 Hz or over 3 Hz.

3.2. Abnormal brain imaging

Six patients (18.8%) showed a single abnormality in each of the following features: hippocampal sclerosis (two patients), hippocampal atrophy (one), hippocampal cyst (one), frontal subarachnoid cyst (one) and communicating hydrocephalus (one).

3.3. Diagnostic sensitivity of video-EEG monitoring

The VEM data were available for 51 patients. Twenty-nine (57%) showed more than one clinical seizure. Twenty patients (69%) only showed myoclonic seizures (MS) and six (21%) demonstrated both MS and generalized tonic–clonic (GTC) seizures with or without any preceding myoclonic jerks. Three (10%) showed only GTC seizures. Although no clinical seizure was observed in 22 patients (43%), 16 of them (73%) had typical generalized epileptiform discharges interictally. A slow wave burst alone was observed in two. Overall, we could diagnose 45 (88%) of all subjects immediately after VEM. Their monitored seizures or accompanying ictal EEG or interictal EEG or its combinations help us to reach the conclusion. Twenty patients (39.2%) were diagnosed by typical interictal changes only, 15 (29.4%) by typical ictal plus interictal changes, and 10 (19.6%) by the combination of typical seizures, typical ictal and interictal changes.

3.4. Conventional EEG versus video-EEG

In 15 patients, a routine c-EEG was performed prior to admission for VEM at our hospital. One VEM result was unavailable for review. Therefore, the VEM data was compared with the c-EEG results in 14 patients. Twelve patients underwent c-EEG only once, but two patients received it three times because they showed different findings each time. Of the 14 patients, 10 showed completely normal c-EEG results. Among these, nine and one patient underwent VEM for 24 h and 48 h, respectively. Six showed both ictal and interictal changes that were compatible with generalized epilepsy, and three had only a typical interictal abnormality. Among the patients with a previously normal c-EEG, nine could be confirmed as having JME using VEM.

Four patients underwent VEM, even though their c-EEGs showed some epileptiform discharges (Table 2). Patient 1 complained of lip smacking during seizure, which was usually regarded as part of the temporal lobe epilepsy, as well as myoclonic jerks and generalized seizures. This patient showed focal sharp waves on the left frontal area in the c-EEG. An atypical seizure history and confusing c-EEG results led to the application of VEM. In this patient, GTC seizures preceded by myoclonic jerks involving both arms accompanying generalized rhythmic spike-and-wave patterns were recorded. This patient also showed frequent interictal polyspike-and-wave features. Patient 2, whose VEM data was lost, had inconsistent atypical findings in three sequential c-EEG sessions. Spike-and-wave patterns over both frontal regions, normal results and sharp waves over the right frontal region were observed serially. Another one patient (patient 3) had experienced frequent myoclonic jerks and generalized seizures. The c-EEG revealed generalized spike-and-wave patterns with a focal spike over the right temporofrontal region. A brain MRI scan revealed right hippocampal sclerosis. VEM showed MS in all four extremities, which corresponded to a generalized rhythmic spike-and-wave complex. The interictal EEG showed similar findings. Patient 4 had a history of both generalized seizures and myoclonic jerks and underwent c-EEG three times. The following inconsistent results were obtained: spike-and-wave dominant on the left frontotemporal region, dominant on the right frontal region and finally generalized rhythmic spike-and-wave complexes dominant on the both frontal regions. This patient showed MS mainly involving the right arm but corresponding to a generalized symmetrical spike-and-wave complex.

3.5. Monitoring duration according to ictal semiology

In all patients, the mean duration of monitoring was 1.8 ± 1.1 days (range 1–6). We recorded only MS without any other types of seizure in 20 patients, only GTC seizures in 3 and both types of seizures in 6. Absence seizures were not observed. The average duration of monitoring was 1.8 ± 1.0, 1.7 ± 0.6 and 2.2 ± 1.6 days, respectively. Comparisons between the groups using ANOVA showed no significant differences in the VEM duration. Twenty patients showing MS alone had a mean of 3.8 episodes (range 1–18) excluding one patient who did not have a precise record because more than 30 seizures had been noted. Only GTC type seizures were experienced by three patients (one, two and three episodes). Six patients with both types of seizure had 6.7 episodes of isolated MS and one GTC seizure with the exception of one with two GTC seizures.

Table 2
Atypical cases of juvenile myoclonic epilepsy

	Seizure history	Conventional EEG (location of EDs)	MRI	Video-EEG monitoring		
				Interictal EEG	Ictal EEG	Monitored seizure
Cases with discordant results between conventional EEG and video-EEG						
1	Lip smacking, M, G	Lt. frontal	–	RSW	PSW	M ± followed by G
2	M, G, A	1st: both frontal, 2nd: WNL, 3rd: rt. frontal	WNL	N.A.	N.A.	N.A.
3	M, G	Generalized and rt. temporal	Rt. HS	RSW	RSW	M
4	M, G	1st: lt. frontotemporal, 2nd: rt. frontotemporal, 3rd: both frontotemporal	WNL	RSW	RSW	M
Cases with unusual semiologies						
5	Visual illusion → G, M, A	–	WNL	RSW	RSW or PSW[a]	M ± followed by G
6	Visual illusion → G, M	WNL	WNL	ISW or PSW	RSW	Myoclonic
7	Ictal amaurosis, M, G, A	Slow wave burst	WNL	RSW or PSW	PSW	M ± followed by G
8	M, G	–	WNL	RSW or PSW	PSW[b]	Right version → G
9	M, G	–	WNL	RSW or PSW	RSW[c], PSW	M (rt. arm) M → Rt. version → G
10	M, G	–	–	RSW or PSW	PSW[d]	M → lt. version → G

ED, epileptiform discharge; N.A., data not available; RSW, generalized rhythmic spike-and wave complex; PSW, generalized polyspike-and-wave complex; ISW, generalized isolated spike-and-wave; HS, hippocampal sclerosis. Other abbreviations are noted under Table 1.

[a] Dominant on right frontal region.
[b] With focal rhythmic alpha on left frontal region.
[c] With left frontal dominant pattern.
[d] Dominant evolution of right frontal region.

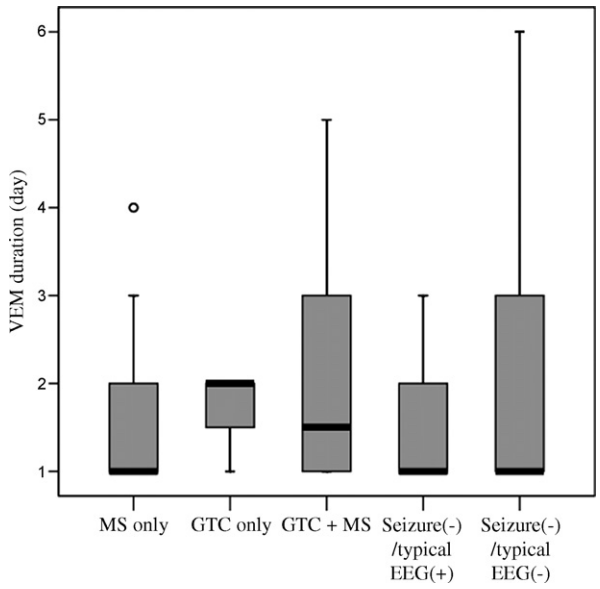

Fig. 1. Duration of video-EEG monitoring (VEM). Patients were divided into five groups; (1) those with only a myoclonic jerk or jerks; (2) those with generalized tonic–clonic seizures (GTC) only; (3) those with both types of seizures; (4) those with no seizures but showing typical interictal EEG changes (5) those with no seizures and without typical abnormalities on EEG. The duration of VEM did not differ significantly between groups. Thick bars in the box denote the median value.

Clinical seizures were not observed in 22 patients. Of those, 16 showed interictal epileptiform discharges that were compatible with generalized epilepsy. Their mean monitoring duration was 1.5 ± 0.6 days. The remaining six patients without any typical EEG changes were monitored for 2.2 ± 2.0 days (Fig. 1).

3.6. Unusual semiologies (Table 2)

Asymmetric episodes of MS were observed in 16 of the 26 patients showing MS during VEM. A right and left dominance was noted in 12 and 4, respectively. Asymmetric MS episodes usually corresponded to symmetric EEG discharges.

Three versive seizures and one circling seizure evolving to GTC seizures were documented in three patients. The ictal EEGs revealed a focal epileptiform activity or dominant evolution pattern in the opposite direction to the versive seizure in two episodes. Patient 8 showed two versive seizures to the right side. Rhythmic alpha waves over the left frontal region evolving to generalized activity were observed at the first episode. However, the next similar versive seizure did not show any lateralized EEG change. Patient 10 had one versive seizure during three GTC seizures. Her left version corresponded to fast spikes that were dominant on the right hemisphere. One symmetric GTC seizure accompanied a preceding isolated focal sharp wave in the left frontal region with subsequent diffuse polyspike-and-wave features. Inconsistently, the other symmetric GTC seizure did not exhibit any focal or asymmetric epileptiform discharge. However, these patients also showed frequent symmetric generalized epileptiform discharges in most parts of their EEG during VEM.

Intriguingly, another atypical history was taken from three patients (patient 5, 6, 7). They had experienced visual aura independently or prior to a GTC seizure, which was considered to be partial epilepsy, as well as myoclonic jerks and absence. Monitoring revealed several episodes of symmetric myoclonic jerks as well as GTC seizures. The EEG finding was also typical for JME, showing symmetrical generalized polyspike-and-waves or generalized spike-and-wave complexes, both interictally and ictally.

Table 3
Current antiepileptic medications and outcomes for 55 juvenile myoclonic epilepsy patients

Current regimen	No. of patients (%)	Seizure free, *n* (%)	>50% reduction, *n* (%)
VPA	18 (33)	13 (72)	4 (22)
VPA + TPM	8 (15)	2 (25)	5 (63)
VPA + LMT	9 (16)	5 (56)	2 (22)
VPA + TPM + LMT	5 (9)	2 (40)	3 (60)
LMT	4 (7)	2 (50)	2 (50)
TPM	6 (11)	3 (50)	3 (50)
LMT + TPM	1 (2)	0 (0)	1 (100)
ZNS	1 (2)	1 (100)	0 (0)
LMT + CNZ + TPM	1 (2)	1 (100)	0 (0)
No medication	2 (4)	0 (0)	0 (0)
Total	55 (100)	29 (53)	20 (36)

VPA, valproate; TPM, topiramate; LMT, lamotrigine; ZNS, zonisamide; CNZ, Clonazepam.

3.7. Patient treatment decisions and outcome

We could categorize treatment decisions after VEM into three groups: (1) changing in-effective or contraindicated drugs, which had been prescribed by the referring physicians ($n = 20$, 36%), (2) *de novo* medication ($n = 17$, 31%), and (3) the adjustment of previous medication, dose-up or add-on ($n = 18$, 33%). In the first group, 15 had previously taken carbamazepine and three oxcarbazepine. After appropriate changes from carbamazepine or oxcarbazepine at discharge, 73% (11/15), 67% (2/3) showed seizure freedom at last follow-up, respectively. Seizure frequency also decreased, more than 50%, in the remaining patients, for both types of seizures, MS and GTC. Among patients with *de novo* medication (second group), 10 patients maintained monotherapy. Nine of those had a same drug prescribed at discharge and one had another drug due to weight gain by valproate. *De novo* monotherapy became polytheraphy in seven patients due to unsatisfactory response of monotherapy during follow-up. Seizure freedom was achieved in six patients on monotherapy and four on polytheraphy. After adjustment of previous medication(s) (third group), seizure freedom were in 8 of 18 patients and more than 50% reduction were in nine patients. Table 3 summarizes the medications currently used by our patients and their seizure outcomes.

4. Discussion

VEM is widely used to identify epileptic syndromes by analyzing the seizure semiology and accompanying ictal or interictal EEG abnormalities.[17] This study investigated patients with JME undergoing VEM and evaluated their clinical details.

Most patients in this series had commonly experienced at least two types of seizure, including generalized seizure among the three representative semiologies of JME. The distribution of the semiological history was similar to that in other studies.[1,11,18] Although approximately 20% of patients were supposed to have experienced absence seizures, they were not detected by VEM. It is believed that a loss of consciousness might be experienced briefly during the period of "interictal" EEG changes, which can be easily overlooked by the patient. Further, it used to be that absence seizures in JME is mild, so it is difficult to detect it, compared to typical absence seizure of childhood absence epilepsy.[19] In contrast, the rate of patients experiencing generalized seizures was higher than other studies,[1,12,20] which is probably because of our study design for selecting the population undergoing VEM.

It is generally accepted that a normal EEG result can delay a correct diagnosis, particularly for patients with an atypical

semiology. Overall, more than 40% of all epilepsy patients had one normal interictal EEG.[21] In this series, the rate of normal EEGs by VEM (11.8%) was much lower than that observed in routine c-EEGs.[1,11–13] In other words, among the 51 patients who were finally established as having JME, 88% had their diagnosis confirmed using VEM. Recently, one study examined the sensitivity of c-EEG performed serially in patients with generalized epilepsies.[22] They reported that 44% of patients in a subgroup with JME showed a normal first EEG, and 17.5% persistently showed normal EEGs, even when repeated four or eight times. Our result was slightly better than that obtained from repetitive EEGs, and it is much worthwhile that the VEM was completed in only about 2 days. Waiting for a typical abnormal EEG finding to appear may thus be time consuming.

There are several plausible explanations for the relatively high sensitivity of diagnosing JME by VEM. Sleep deprivation during monitoring is one possible reason because it is a precipitating factor in all epilepsy syndromes,[23,24] particularly in JME where it increases the interictal epileptiform discharges and habitual seizures.[25–28] Although the sleeping status was not verified in this series of patients, self-induced sleep deprivation might have contributed to the high sensitivity of the EEG results. In one report,[27] a sleep-deprived EEG for approximately 30 min increased the sensitivity of generalized epileptiform discharges in JME patients from 70.3% to 86.5%, which is similar to our results. Although EEGs performed on sleep-deprived patients with JME can increase the yield of generalized epileptiform discharges, this remains a worrying problem because it might induce seizures in outpatient EEG laboratories. VEM can be superior to c-EEG with the safe monitoring of seizures in an inpatient setting. In addition, the drug-free state of our patients during VEM might have contributed to our high diagnostic sensitivity because the EEG results may be entirely normal in patients with the appropriate drug treatments. JME is usually characterized by myoclonic jerks in the morning, and more than a half of patients with JME experience seizures upon awakening.[12] Considering that accompanying EEG abnormalities are also apparent in the morning, EEG monitoring that includes this time window should be given particular emphasis as one possible explanation. One recent prospective study also established the importance of morning EEGs in the diagnosis of JME.[29]

Another point of interest is that 9 of the 10 patients with previous normal "routine" c-EEGs were diagnosed at the time of VEM. The video recordings revealed five patients to have MS only and one patient to have several episodes of MS and one GTC seizure. Three of the remaining four patients did not show any clinical seizures but their video-EEG revealed abnormalities compatible with generalized epilepsy. Seizures and EEG changes could be detected in 90%, even when the patients showed normal results in one or more c-EEGs before VEM. This is similar to the overall rate of diagnosing JME through VEM as well from serial c-EEGs. Taken together, we can deduce that an early VEM would have been more relevant for making an early diagnosis in these patients.

Although it has been considered classically that no structural lesion is observed in patients with JME, according to recent reports, MRI abnormalities occur in 16% to 26.8% of such patients. In this study, 6 out of 32 (19%) patients showed abnormalities on brain imaging, which in agreement with other prospective series.[30,31] Patients who complain of a history of unidentified seizures with a hippocampal abnormality are often considered to have partial epilepsy originating from that abnormality. Although three of our patients with hippocampal abnormalities had a tentative diagnosis of JME based on the history of myoclonic jerks and generalized seizures, the c-EEGs could not support their clinical history. However, there was one patient (patient 3) with generalized as well as focal spike-and-wave features on the ipsilateral side of the hippocampal lesion on c-EEG. Thus, VEM clarified the diagnosis, showing symmetric MSs of all four extremities in association with the generalized epileptiform discharges. Despite not detecting any clinical seizure in another patient during monitoring, fortunately we found generalized poly or isolated spike-and-wave complexes interictally. In the other patient with asymmetric hippocampi, we could record MSs with symmetric involvement of both arms. Therefore, the VEM results can reinforce a diagnosis of JME based on the clinical history. Some authors reported[32] that comorbid patients having both partial and generalized epilepsy were present in 0.2%. Their focal lesions always revealed hippocampal sclerosis. However, our patients were different from their patients, who invariably had a symptomatic hippocampal abnormality.

Another intriguing semiology was found in the clinical history. Visual aura was present in three patients. In two of them, visual illusions preceded the GTC seizures and the other patient complained of ictal amaurosis. The presence of a myoclonic component in both the clinical history and VEM suggests JME rather than occipital lobe epilepsy. It could not be verified whether or not the two syndromes coexisted because VEM had been performed for a relatively short duration. In one study,[33] the authors suggested a genetic overlap between idiopathic occipital lobe epilepsy and JME because one patient showed the typical semiology of JME and visual aura. In addition, there were two syndromes in different members of a family, which suggested that a single disease had different phenotypes. However, this could not be confirmed because the family history was not examined in this series. It was presumed that more patients with JME experience visual symptoms than expected.

Asymmetric semiological features, such as the asymmetric MS and the asymmetric evolution of GTC seizures were also observed in our patients, as shown by previous studies.[1,5] The higher rate may have been biased by the selection of patients for admission to VEM, applied more in patients with an atypical history. Of interest, it was found that a unilateral dominancy of MS did not always appear and the dominant side did not change during monitoring, which is in contrast to another study.[15] The criteria for diagnosing the unilateral nature of a MS may account for this discrepancy.

Although a versive seizure (VS) or circling seizure (CS) can occur in patients with partial epilepsy syndrome, particularly in those with frontal lobe epilepsy, the presence of these was confirmed in the patients with JME, as in other studies.[34,35] The direction of VS or CS appeared to be towards the contralateral side of the dominant ictal EEG findings, as was observed in two of the four episodes of VS or CS. This observation is supported by a previous report.[5] However, it is still unclear whether a generalized epilepsy syndrome has laterality on the ictal EEGs when a patient shows such seizures.[6] In this study, because VEM captured multiple symmetrical generalized interictal epileptiform discharges in most cases, it avoided confusing generalized epilepsy syndrome with focal epilepsy despite the incidental focal or lateralized EEG features. Anecdotal reports on this issue still have not presented a consistent finding. Therefore, more cases will be needed to clarify it. As with asymmetric MSs, GTCs were not always accompanied by a VS or a CS in our patients, which is in agreement with other series.[21,35] To date, it has been assumed that focal or lateralized features arise from regional hyperexcitability and focal microdysgenesis.[36] Unstable regional hyperexcitability might influence such inconsistent laterality. We consider that the asymmetry of the semiology in JME as well as that of the ictal or interictal EEGs is probably a temporary phenomenon.

Meanwhile, in terms of outcome, many studies have reported that 64% to 95% of JME patients showed the good response to individual antiepileptic drugs.[37–40] In our study, overall, approximately 90% of patients achieved good outcomes (53%, seizure free

and 36%, >50% seizure reduction) at last follow-up, in agreement with other studies.

The clinical value of VEM in revealing a patient's habitual seizure is crucial because both the patients and their witnesses are uncertain of their semiology and neurologists are either not always convinced of an atypical history of seizures in patients with JME or have overconfidence in it. Although it seems to be reasonable to apply the alternative methods such as morning or sleep-deprived EEG for putative JME patients with classical semiology, it is different in cases of atypical semiology or inconsistent clinical results. We suggested that VEM could be more relevant for patients showing a discrepancy between the c-EEG results and their historical semiology, e.g., those showing asymmetric seizures with a normal EEG and visual aura with generalized epileptiform EEG changes. This is also necessary for patients with a focal abnormality alone or lateralized abnormality shifting its side within one c-EEG recording. Because the focal or atypical lateralized findings of EEG or semiology do not appear consistently, we propose that a more comprehensive review of interictal EEGs over a long duration using VEM system would be helpful. Of course, since VEM is used to verify a vague semiology, it can reveal easily overlooked MS or absence seizures in patients with JME. The elicitation of two semiologies of JME together with generalized seizures by VEM leads to a confirmation of JME among several categories of generalized epilepsy syndrome.

In summary, VEM for a relatively short duration can be useful and feasible for making a diagnosis of JME. First, VEM can detect the EEG changes and seizures of patients with JME more easily by covering the whole circadian cycle as well as applying provocation methods, such as sleep deprivation and AED withdrawal. Second, the longer session of EEG than that of c-EEG makes it possible to detect many symmetric EEG abnormalities even in the presence of infrequent focal abnormalities. Thus, VEM is a time-effective mode in arriving at a diagnosis of JME. The main limitation of this study was that patients with well-controlled seizures, who were referred from other clinics and had a definite diagnosis of JME both clinically and electrophysiologically, did not undergo VEM and therefore were not included in this study. However, in terms of cost effectiveness we still question whether VEM is superior to repetitive conventional EEGs or sleep-deprived EEG for a diagnosis of JME. Further prospective controlled evaluations are required to clarify this.

Conflicts of interest

There are no conflicts of interest in this study.

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